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Answer to Case of the Month #158 Lymphocytic Interstitial Pneumonia

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Clinical Presentation

A 52-year-old woman with a 10-year history of seropositive rheumatoid arthritis presented with insidious onset of breathlessness over a 6-month period. Clinical examination was unremarkable. She was acyanotic, not clubbed and on auscultation, there was good air entry bilaterally and no crepitations. Pulmonary function tests revealed a mild restrictive defect and reduced transfer factor. Chest radiograph showed haziness in the perihilar regions, with widespread nodularity. A high-resolution computed tomography was performed (Figure 1).

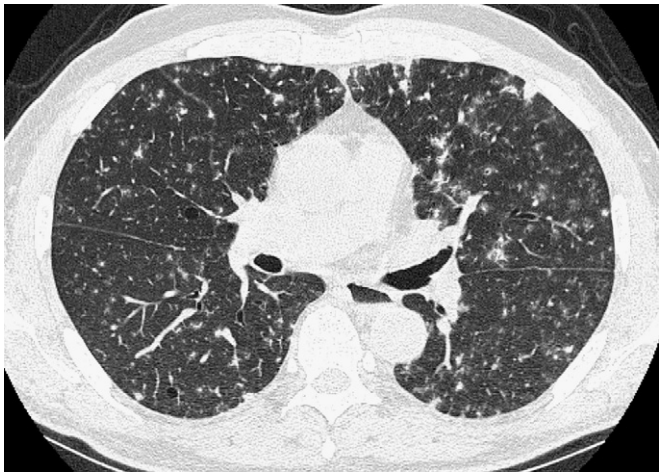

Figure 1. High-resolution computed tomography at the level of the main pulmonary artery, showing multiple ill-defined pulmonary nodules. Two thin-walled cysts are seen in the right lung.

Diagnosis

Lymphocytic interstitial pneumonia (LIP).

Discussion

LIP is a benign lymphoproliferative disorder characterized by a diffuse interstitial proliferation of small lymphocytes and plasma cells [1]. It is associated with a variety of conditions, including dysproteinaemia, autoimmune disorders, collagen vascular diseases, and AIDS. It is rare in human immunodeficiency virus–infected adults, but it is one of the defining criteria for AIDS in children under the age of 13 years. The most common symptoms are gradual onset of cough and dyspnoea over a period of months or years. Pulmonary function tests usually reveal a restrictive ventilatory defect with a low transfer factor. The chest radiographic appearances of LIP are nonspecific, and high-resolution computed tomography (CT) is the modality of choice for evaluation and guiding lung biopsy. Characteristic CT findings include multiple ill-defined centrilobular nodules, areas of ground-glass attenuation, and thin-walled cystic airspaces [2]. The major differential diagnoses are low-grade lymphoma and some infections that can cause a similar appearance, for example, *Pneumocystis carinii* pneumonia. Immunohistochemical analysis, molecular gene rearrangement studies, and silver staining are often required to exclude these alternative diagnoses [3]. In the past, LIP was considered a risk factor for development of malignant lymphoma, however, many previously described cases were reclassified as lymphoma from the outset, and only a small number of definite LIP cases actually undergo malignant transformation. The natural history of LIP is highly variable; although some patients improve after treatment with corticosteroids, more than one-third have progressive disease.

Key Words: Computed tomography; Interstitial pneumonia.

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